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Report of a Case of Osteitis Deformans

BY

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Report of a Case of Osteitis Deformans.

By Charles Herwirsch, Ph. M., M. D.

ON FEBRUARY 3d, 1896, my attention was called to an inmate of the Nazarene Home for Aged People, of this City, of which institution I am House Physician.

The patient, K. L., a single woman, 71 years old and about 5 feet in height, was sitting in a large chair with her feet crossed. Her head, which was of unusual size, was bent forward, her lower lip hung down and saliva was dripping from her mouth. The superficial blood-vessels of the head were prominent and pulsating. The hair, which had been cut short, was divided above the very broad and prominent forehead and hung down on both sides, leaving a large bald place on the vertex and occiput.

The face was small, the whole head was rounded and the appearance was that of spurious hydrocephalus. The hands and feet were normal in size, the ankles edematous. A chest examination disclosed a fairly normal condition; an accentuated second aortic sound was observed, with a faint blowing murmur in the neck. When interrogated, the patient could tell the year of her birth and would give tolerably rational answers to simple questions; but she was very selfish, and if not attended to at once, when necessity required, she would scream, cry murder etc. The appetite was ravenous at the time of my first visit and the patient suffered with a profuse diarrhea, having from ten to twelve stools a day. From her brother I learned that K. L. enjoyed good health up to her 64th year; she was a normal individual, mentally and physically, of a very lively disposition and a hard worker. When she arrived at that age she was attacked with rheumatism and confined to bed for one year, when the gradual enlargement of the head began. She was placed in charge of a poor family who reported about three years ago that she was, as they described her condition, paralyzed.

Until a few months ago she was able to go about her room with the aid of a cane, but in January of the present year, when she was admitted to the home where I found her, she was unable to walk unattended and had to be helped in and out of bed, by one and later on by two persons, on account of her great weight and feebleness. While under observation she generally remained quiet during the day, sitting in her arm-chair, usually listless, never conversing, making only short replies when spoken to. She was able to recognize me, the matron or her nurse; she never complained of pain but she did not like us to palpate her head. The pulse was strong, 70 per minute, and regular; respirations normal. The urine was voided in bed, on account of the helpless condition of the patient; its specific gravity was 1014 and it contained some albumin; on account of the difficulty of procuring another specimen I was unable to make more than one examination.

The bowels were easily controlled by regulating the diet and the administration of opium and lead acetate for a few days ; and up to the time of her death, March 13, she never had more than two stools daily. I had much more difficulty to control her temper and insomnia. She was very restless and troublesome at night, frequently waking the other inmates by her cries, if her wants were not immediately supplied. I gave her in succession chloral, bromids, sulfonal, trional paraldehyd, and morphin hypodermatically, but these drugs would quiet her only a short time ; I then prescribed a combination of 30 grains of bromids, 15 grains of chloral hydrate and ¼ grain of morphin, which would have the desired effect when given at 9 and 12 p. m. Infusion of digitalis somewhat modified the edema of the ankles and decreased the frequency of urination.

The condition grew gradually worse, the patient taking less interest in her surroundings ; on the 10th of March she refused to take food ; on the 11th she became comatose and died in coma on the 13th.

The accompanying photographs were made by Dr. Cuvier R. Marshall. Dr. Samuel Wolfe had the kindness to see the patient with me on several occasions and made the following notes :

The patient was seen Feb. 28th 1896. The head measured 28 inches fronto-occipital circumference, from the meatus auditorius of one side to that on the other, and over the vertex, 17 inches. The enlargement is perfectly symmetrical and the sutures are not separated. The veins are very prominent over the scalp, the hair is thin. The patient sits with the head flexed on the chest, the eyes cast down, the arms and legs crossed, and presents a listless and apathetic appearance. With the hands placed on the head, there is felt a pulsation in all parts of the scalp, slightly stronger on the right half than on the left: through the stethoscope placed on any part of the scalp, there are heard an inspiratory and an expiratory sound of somewhat higher pitch than the corresponding sounds heard over the sternum. These transmitted sounds are accounted for by the bone changes found in the thorax and skull. The voice is transmitted to the skull as a vocal fremitus.

The intellect, though dull, is not entirely destroyed. Her answers to questions are often correct, and when erroneous she makes efforts, though not always effectual, to correct herself. She is delirious, and sometimes bellicose and abusive to her nurse, at night. She voids urine and feces in the bed, but will use a vessel if her attention is directed to it. She has therefore not a true incontinence, but is simply apathetic. The left pupil is myotic, the right normal and mobile.

When seen last, March 12th, 24 hours before death, the right pupil was dilated, making the inequality greater than previously. She was comatose ; the respiration was blowing, and the right side apparently paralytic. The pulse was 140.

The autopsy was performed by Dr. Frank Savary Pearce, in the presence of Drs. Samuel Wolfe, J. P. Crozier Griffith, Wm. E. Robertson, H. B. Nightingale, Victor T. Roth, and myself, with the following result : Post

mortem rigidity was well marked ; there were no scars or signs of abnormal growth except the greatly hypertrophied calvaria, and the slight bowing of the long bones of the lower extremities, most marked in the left leg, as observed by Dr. Griffith. The lower half of the head and face was not enlarged, and the fingers and toes presented none of the spade-like appearance of acromegalia. The lips were prolapsed and a little thickened, but there was no true myxedema. The thyreoid gland was not palpably enlarged ; the skin everywhere had a muddy appearance, characteristic of the prolonged defective nutrition of the body, generally. The temporal arteries were tortuous and dilated, and the veins over the scalp were somewhat distended ; the hair on the scalp was very sparse and rather coarse.

On making the scalp incision evidences of venous engorgement were apparent, although the skin was not thickened. The bony structure of the skull was unusually soft and friable, the saw penetrating most readily ; it was at once noticed that the thickness of bone was great and that there was little diploe apparently. The density was greatest in the temporal regions. On final removal, it was found that the vault of the skull measured 1⅜ to 1½ inch in thickness in the occipital region, and ¾ inch in the frontal region, varying between these limits at intermediate points. The corrected measurements of the skull cap, on the plane of the incision are as follows : Thickness in occipital region, 1⅜ inch ; frontal, ¾ inch ; long diameter, external, 9 inches ; internal, 7 inches ; short diameter, external, 7 7-16 inches, internal 5 10-16 inches.

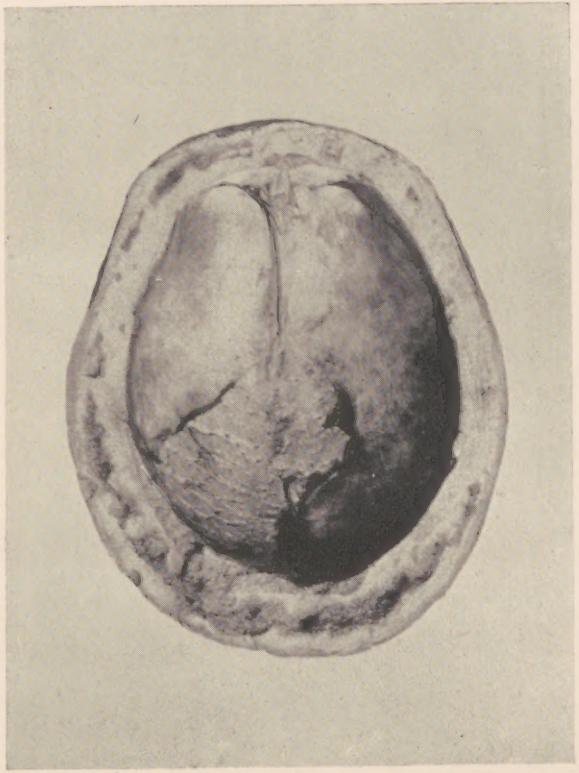
The bone substance could be easily pricked out with the scalpel and in the temporal regions there was an absolute fusing of the once calcareous bone of the internal and external plates ; while in the remaining circumference it was apparent that the relative amount of diploe was much diminished, in proportion to the greatly increased size of the bone.

In fact little diploe may be seen except in the occipital bone ,the thickest portion of the skull. The basilar process of the occipital, the body of the sphenoid and the other bones forming the base, were very easily penetrated by the chisel. There was nothing in the brain membranes having a pathological bearing, except that the dura was slightly indurated and thickened.

The detached skull cap weighed 4 pounds, 2 ounces, the brain weighed 48 ounces, was not enlarged and fitted nicely in the normal sized cavity of the vault.

The ventricles were distended beyond the normal with clear serum : the ependyma was normal ; no hemorrhagic lesion or other alteration was found in the encephalon. The head enlargement was, therefore, entirely in the bony structure, and in order to determine what might be the nature of the process it was clear that further examination of other bones was necessary.

Both femora were found to be thickened and slightly curved, with the convexities forward and outward. The epiphyses were much enlarged and covered with a new growth, capped, as it were, with osteoplastic incrustation (exostosis) of similar friable, mealy bone, more so than was exhibited



DR. HERWIRSCH'S CASE OF OSTEITIS DEFORMANS.

in the skull cap, where the densest bone of the body was found. The left tibia was greatly thickened and curved forward and outward. The medullary cavity was disproportionately enlarged and filled with red marrow, some of which, with the adjacent bone, was removed for microscopic study. The right tibia was little affected. The bones of the arms, hands and feet were not noticably enlarged, with the exception of a slight protrusion and thickening of the calcanei, posteriorly.

Median abdominal incision revealed much yellow fat beneath the skin and in the omentum. The ribs also proved to be soft and friable, and were readily incised with the ordinary scalpel used in the section. The bone marrow was of a fatty consistence, macroscopically. The crests of the ilia were more blunted and larger than normal. The same was true of the os pubis. Portions of these bones were removed for further study.

The pleural and pericardial sacs contained normal quantities of fluid, and were non-adherent throughout. Aside from slight emphysematous areas anteriorly and above, and post-mortem congestion at the bases, behind, both lungs presented no pathological changes.

The heart was fatty and slightly hypertrophied. There was a very large and dense chicken-fat clot extending into the aorta from the left ventricle, and a similar clot adhered to the mitral leaflets and their cordæ tendinæ, demonstrating the slow mode of death.

Atheroma existed at the base of the posterior mitral leaflet. The mitral orifice admitted two fingers readily. In the right heart much white clot was also found, and less currant-jelly clot ; the tricuspid opening readily admitted the passage of three fingers. The wall of the right ventricle contained a larger proportion of fat than of muscular tissue. The liver was normal in size, and was the seat of a slight cirrhosis and fatty degeneration. The kidneys were slightly enlarged, the cortices were somewhat narrowed, and the capsules were stripped off readily, although beginning interstitial change was evident. The spleen was slightly under size, while the intestines and other abdominal viscera were normal.

I quote from Dr. Pearce's report before the Pathological Society the following : "Just where this case belongs in pathology it is not so easy to determine. The name, rarefying osteitis deformans, suits for most of the bony changes, for there is certainly porosis with deformity. Slides of bone from the pubes and femur, made by Dr. Robertson, show absence of bone cells, and irregularly enlarged Haversian systems, the evidence of chronic inflammation. The sections of bone and marrow from the left tibia, kindly made for me by Dr. A. E. Taylor, show a queer lot of changes, viz. : Atrophy of bone ; a great loss in number of osteophytes and osteoblasts ; conversion of parts of the bone into mere fibrous structure ; the marrow contains very few myelocytes. It would seem that the changes here found indicate an advanced state of this interspersed skeletal disease, in which detritus was the remnant. The bone was decalcified in 24 hours (with 2 per cent. HCl in 70 per cent. alcohol), while in the case of normal bone a week would be required.

What then is this unique bone affection? Rickets is, of course, out of court, as this disease in which curved bones are common, never furnishes such enormous enlargement, and it occurs in children. In mollities ossium the bones are elastic, therefore not friable, sclerosed or brittle. In acromegalia the lower portion of the head mainly is enlarged, and not the calvaria: the jaws are somewhat prominent, the hands present marked evidence of bony growth, and the pituitary body would likely have been affected. In leontiasis ossium, one would expect firmer sclerosis than is presented in this skull cap which is most dense, and yet less than normal bone, with all its thickness; the malar prominences would have been more conspicuous and, as inferred, even eburnation predominates, not friability. May not leontiasis, after all, be the further condensing osteitis?

This extreme bony enlargement is a very rare occurrence and its association with wide-spread bone disease I cannot find recorded anywhere. The weight of evidence seems to be in favor of a chronic rarefying osteitis with deformities, hypertrophy, and a final condensation as shown in the skull."

In addition to Dr. Pearce's clever differentiation of the various bone diseases which should be considered in connection with this case, I wish to present a description of osteitis deformans as outlined by Dr. Paget, of London, on November 24th, 1876, when the first case on record was reported.

Osteitis deformans commences after middle age, is very slow in its course, and it progresses without affecting the general health, determining no other troubles than those which result from alterations of the shape, volume and direction of the diseased bones. Even when the cranium is enormously thickened and when all the bones are extremely altered in their structure, the intelligence remains intact.

The disease usually affects, in the first place, the long bones of the lower limbs and those of the cranium. It is usually symmetrical; the bones increase in size and soften; the affected portions of the skeleton which bear the weight of the body are bent and deformed by the abnormal curvatures which they assume. The natural curves of the vertebral column increase and thus results a considerable diminution of the height of the patient, which has already undergone a considerable reduction on account of the bending of the diaphyses of the tibiae and femora and the lowering of the heads of the latter. It is an important point that this softening, this loss of resistance is accompanied by neither weakness of the limbs nor fracture.

The legs remain strong and in condition to support the weight of the body: they bend, but do not break. In the first stages, or sometimes from beginning to end, these patients have pain in the affected bones. They are taken to be cases of rheumatism, of chronic periostitis, syphilis, gout, neuralgia, etc.; there never is fever nor any abnormality of the urine. There is no association with syphilis or any other constitutional disease except cancer.

In the first case of Paget there were cancerous nodules in the walls of the cranium. There was a peculiar inflammation affecting the cranium and,

in the long bones, chiefly the compact layer of the walls of the diaphyses as as well as that of their articular surfaces. It is probable that these changes were inflammatory from the beginning, because the softening was accompanied by thickening, by proliferation of imperfectly formed bone and by hyperemia..

The microscopical examination of Dr. Paget's first case, made by Mr. Butin, seems to indicate that this affection should be classed with true osteomalacia as a benign and hypertrophic variety. C. N. Macnamara, in his work on diseases of the bones and joints, comes to the conclusion that osteitis deformans is usually the result of a chronic rheumatic osteitis.

Since Paget described his first case, in 1876, only a limited number of cases of this disease have been reported ; the list is appended as follows :

S. W. Wheaton ; Ivory Exostosis of the Skull. Read at the London Pathological Society in 1892.
A. Watson ; A Case of Osteitis Deformans. Melbourne, 1894.
W. Dubreuilh, in *Arch. Chlin. de Bordeaux*, iv., 1895.
L. H. Guthrie ; *Trans. Med. Soc.*, London.
T. Bryant ; *Guy's Hospital Report*, London, 1877.
C. N. Ellinwood ; *Western Lancet*, San Francisco, 1883.
I. F. Goodhart ; *British Medical Journal*, London, 1888.
L. Guinon ; *Bul. Soc. Anat. de Paris*, 1885.
J. R. Lunn ; *St. Thomas' Hosp. Report*, London, 1883 and 1884.
The same writer ; *Med. Times and Gazette*, London, 1885.
The same writer ; Four Cases in *Transac. Chlin. Soc.*, London, 1885.
McShedran ; *Medical News*, Philadelphia, 1885.
Martel ; *Gazette Med. de Paris*, 1886.
H. Morris, *Transac. Path. Soc* , London, 1882.
J. Paget ; *Med. and Chir. Transac.*, London, 1877.
Pick ; *Lancet*, London, 1883.
Power ; Two cases Path Society, London, 1886.
Pozzi ; in Paris, 1885.
B. Smith, Leipzig, 1884.
A. Q. Silcock ; Pathological Society, London, 1884.
The same writer, with specimens taken from a woman aged 82, *British Med. Jour.* London, 1885.
F. A. Southern, *Med. Chron.*, Manchester.
C. J. Symonds ; *Guy's Hospital Report*, London, 1881.
H. L. Taylor, Report of Two Cases, *Trans. American Orthopedic Association*, 1892.
G. Thibierge ; *Bul. M. S. Hop. de Paris*, 1893
Alphonzo reported a case in 1893.
H. Cretier ; Observations on a Case, 1890.
V. P. Gibney ; *Med. Record*, New York, 1890.
W. Edmunds ; Affecting a Single Bone, *Med. Press and Circular*, London, 1890.
I. Mackenzie ; An Arrested Case of Osteitis Deformans, in which fracture of the affected bone took place, *Medical Press and Circular*, London, 1890.
H. H. Clutton ; Chronic Osteitis of the Tibia. (Osteitis Deformans?) *British Med. Journal*, London, 1861.
H. Meunier ; A Case in Paris, 1894.

Osteitis deformans is a rare disease, as may be seen by the small number of cases reported. This case, reported by myself, presents, very likely, the thickest skull cap on record, viz :—1⅜ inch. There is a specimen about one inch in thickness in the museum of the Medical and Surgical Society, London, and another in the museum of the Harvard Medical School, of about the same thickness.

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